

# **Proposed BBUGSS Guidelines on Achalasia**

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## **Definition**

The term 'achalasia' is derived from the Greek, *chhalasis (relaxation)* to describe a condition where there is failure of the gastro-oesophageal junction (GOJ) to relax.

## **History**

**Thomas Willis (1679):** *'I prepared an instrument for him like a rod of a whale bone with a little round button of sponge fixed to the top...'*. The patient underwent a series of dilatations (1).

Achalasia was thought to be spasm of the GOJ for a long time:

1888 Max Einhorn (USA) and Meltzer (Germany) suggested spasm theory incorrect

1904 Gottstein suggested doing a Ramstedts type procedure

1908 Plummer performed a balloon dilatation

1913 Ernst Heller (1877-1964) planned to do a cardioplasty but the oesophagus was thick and difficult *'it was as big as the chimney of a kerosene lamp'*, so he did an anterior myotomy of 8cm, which he covered with a strip of netting and did the same posteriorly. The patient was well 8 years later (2).

1915 Perry (US) and Hurst (UK) termed 'achalasia' and treatment options were:

- a) Forceful retrograde dilatation (through a gastrotomy) – Mikulicz- Schloffer
- b) Oesophagostomy (Zaaijer)
- c) Oesophagogastrostomy (Heyrovsky)
- d) Cardioplasty (Wendel)
- e) Myotomy (Heller)

1918 Berg (US) took up cardiomyotomy

1920 Rowlands (UK) did similar

1991 Pinotti – performed the first laparoscopic Hellers (3).

## **Epidemiology**

Equal sex distribution

Incidence 1/100,000 year<sup>-1</sup>

Prevalance 10/100,000

Peak incidence 30-60 years of age (4)

Chronicity affects patients health related quality of life, work productivity and functional status

Increased rate of SCC oesophagus (3.5%) (4)

## Pathophysiology

### **Primary achalasia**

Characterised by progressive loss of ganglions of the myenteric plexus or idiopathic aetiology. Selectively, there is more loss of the inhibitory nitrergic neurotransmitters. There may be circumstantial evidence of viral exposure and auto-immune responses but not invariably(5). It is worth noting that there is an increased risk of oesophageal cancer subsequent to achalasia.

### **Secondary achalasia**

Associated with protozoan infection with *Trypanosoma cruzi*, secondary achalasia may arise in what is otherwise known as Chagas disease. Achalasia secondary to Chagas can be treated in the same manner as idiopathic achalasia.

Secondary achalasia may also be associated post operatively in fundoplication procedures. *Pseudo-achalasia*s; may arise secondary to cancers, either directly or occasionally in a para-neoplastic fashion.

## Diagnosis

Usually middle aged

Dysphagia affecting solids and fluids

As it progresses may get regurgitation and overspill lung issues

Chest pain occurs – more with vigorous spasm

Assess lungs and nutritional status

### Symptom severity:

The Eckardt score (6) uses 4 symptoms (dysphagia, regurgitation, retrosternal pain and weight loss) to assess achalasia outcomes and, although not validated can be used as an assessment of initial and follow up symptom severity.

Score	Weight loss	Dysphagia	Pain	regurgitation
0	None	None	None	None
1	<5kg	Occasional	Occasional	Occasional
2	5-10Kg	Daily	Daily	Daily
3	>10Kg	Each meal	Each meal	Each meal

Score is 0-12 at baseline

Follow up scores <3 represent a good outcome

## Investigations

### OGD

Tight non-relaxing junction  
Residue in oesophagus  
Should be performed to exclude malignancy (7)  
Not diagnostic alone

### Barium / Contrast

Dilatation  
Hold up  
Disordered peristalsis  
Tapering – ‘birds beak’  
Not diagnostic alone

A timed barium swallow (where the patient drinks 100-200mls of low density barium over 1 minute and films are taken at fixed time points to allow inter-observer consistency) can aid diagnosis and predict response to treatment (7)

### Manometry

Gold standard  
Hypertensive LOS (normal 28mmHg)  
Failure of LOS relaxation  
Aperistaltic oesophageal body  
Raised resting oesophageal pressure

## Classification:

The Chicago Classification of oesophageal motility (v3) is clinically relevant. It prioritises (sequentially) (8):

1. Disorders of outflow of GOJ
  - a. Characterised by a raised IRP (integrated relaxation pressure >15mmHg)
2. Other major disorders of peristalsis – absent contractility, distal oesophageal spasm and ‘jackhammer’ oesophagus
3. Minor disorders of peristalsis – ineffective motility and fragmented peristalsis

Under ‘1. disorders of outflow obstruction’ (cf Achalasia);

Type I: No contractility  
Type II: No contractility and panoesophageal pressurisation in >20% of swallows  
Type III: No contractility and > 20% spastic contractions with DCI (Distal contractile integral) >450mmHg

## Treatment

### Medical

Nitrates, phosphodiesterase inhibitors and calcium channel blockade have been used, but lack convincing evidence for their benefit. Hence, there is no reliable drug therapy (5).

### Endoscopic

#### 1. Pharmaceutical

Injection of 100iu of **botox** into the LOS – temporary benefit 4-6 months. Therefore not recommended in those fit for more definitive treatments. Can be used for those who are unfit or as a bridge to other treatments. Repeat treatments seem to decrease the efficacy.

#### 2. Mechanical

Progressive **dilatation** 30-35-40mm decreases the likelihood of perforation. TTS balloons give radial force and not shearing longitudinal force. Main complication is perforation (gastrograffin swallow is the recommended investigation to exclude a perforation if it is suspected) Beneficial in 7-90% at 1 year  
4-40% experience GORD afterwards  
Suitable in type I & II (9)

There is little evidence of benefit for endoscopic **stent** placement as a treatment for achalasia

#### 3. POEM

Medium term results are similar to surgery or dilatation, in terms of swallowing but there is a higher incidence of GORD. Can be used despite previous dilatation or botox and if symptoms persist after surgery. Best reserved for type III (9)

### Surgical

#### 1. Cardiomyotomy

Best outcomes are for types I and II

Length is debatable – proximally can be limited to endoscopic visualisation that the obstructing segment has opened up. Distally onto stomach, at least 1 cm and up to 3cm but longer incision may increase reflux. Current recommendations is for at least 6cm proximally and 2cm onto stomach.

Addition of partial fundoplication debatable – 1 small trial showed decrease in post-op reflux from 48-9% with a 'Dor'. Current recommendation is to perform a partial wrap to reduce the risk of reflux

Successful in >80%

More beneficial for dysphagia than balloon dilatation in 2 (underpowered) PRCTs. More GORD. QALYs similar for dilatation and Hellers. Dilatation more cost effective.

Persistent dysphagia can be treated with dilatation

#### 2. Cardioplasty

Stapled across the GOJ or incision through GOJ with fundal patch  
Suggested as a useful tool for resistant cases (10)

### **3. Resection**

Even with a 'sigmoid' oesophagus, First line treatment should be Hellers, graded dilatation or POEM (6).

A small proportion of patients have refractory and 'end stage' achalasia (i.e. that which requires oesophagectomy) – massive dilated and tortuous non-functioning oesophagus. This can be on presentation or subsequent to failed Hellers, Dilatation or POEM. Ideally vagus sparing oesophagectomy should be considered.

#### **Follow up and surveillance**

No clear guidance, but pH testing (objective) for GORD is recommended in collaboration with Eckardt score (subjective). Some centres advocate 3 yearly endoscopic or radiographic surveillance for patients with achalasia for 10-15 years (11).

#### **Recurrent symptoms after treatment:**

Recurrent symptoms or Eckardt score >3 should prompt repeat objective testing, with endoscopy, timed barium swallow and manometry.

After failed balloon dilatation, referral for Hellers or POEM is appropriate  
After failed Hellers or POEM, dilatation is appropriate.

Despite an increased risk of cancer in achalasia patients, there is insufficient evidence for routine endoscopic surveillance after treatment for achalasia (11)

#### **Achalasia in Children**

Should follow the same diagnostic and therapeutic pathways as for adults, with the exceptions that surgery is preferred to dilatation and botox is not appropriate.

#### **Organisation of UK Achalasia treatment centres**

Achalasia is rare. Treatments for it are infrequently used outside of specialist centres and are, on occasion, attended by serious complications. According to the SWORD database, the average number of Hellers per unit is 6 per year and 1-2 per surgeon. There should be a move towards high volume centres that offer all aspects of care (diagnostic and therapeutic), to standardise care and allow comparative studies.

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